International Journal of Surgery 12 (2014) 1291–1294

Contents lists available at ScienceDirect

International Journal of Surgery

journal homepage: www.journal-surgery.net

Original research

Solitary fibrous tumor of the liver: Report of two cases and review of the literature

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HIGHLIGHTS

- Solitary fibrous tumor SFT is a rare neoplasm composed of spindle cells.
- The CD34 antibody is not specific for SFT diagnosis.
- It is difficult to define the evolution, the risk factors and the malignant potential of these tumors.
- Surgical resection remains the optimal treatment.

ARTICLE INFO

Article history:
Received 12 October 2014
Received in revised form
24 October 2014
Accepted 27 October 2014
Available online 4 November 2014

Keywords:
Liver
Solitary fibrous tumor
MRI
Surgery
Histopathology

ABSTRACT

A solitary fibrous tumor (SFT) of the liver is a rare neoplasm of mesenchymal origin. 59 cases have been reported in the literature. We report 2 patients who presented with a hepatic solitary fibrous tumor. The first case is a 65-year-old man who presented with an accidental finding of a large mass in the left liver. Biopsy revealed an SFT and left hepatectomy was performed. The diagnosis was confirmed by histopathology. The second case is an 87-year-old woman who presented with disturbances in her liver function tests. A Computed Tomography (CT) scan showed a large mass in the right liver. Surgery was contraindicated because of the patient's poor general condition. A biopsy was done and SFT was diagnosed histopathologically. SFT are usually benign but the risk of malignant transformation always exists, which mandates surgical resection as the optimal management of these tumors. However, because of the small sample size and the rarity of the entity, it is difficult to define the evolution, the risk factors and the malignant potential of these tumors.

1. Introduction

A solitary fibrous tumor is a rare neoplasm which composes of spindle cells and interspersed collagen. It is usually found in the thoracic cavity and the pleura. Rarely, it is found in the mediastinum, skin, meninges, orbit, upper respiratory tract, breast, thyroid and peritoneum. The hepatic location is extremely rare, and to the best of our knowledge, only 59 cases have been described in the medical literature. Although most SFT have benign behavior, some may have malignant features including metastatic spread and local recurrence [1,2]. The clinical presentation is usually mild and non specific, with some patients being asymptomatic and some presenting with abdominal pain, distension, dyspepsia, and other non specific symptoms like weight loss, nausea and vomiting. The clinical manifestation is affected by the size and the location of the tumor [1–4].

Diagnosis of SFT is difficult on imaging studies because imaging features are non specific and do not differentiate benign from malignant SFT. In addition, other benign and malignant tumors such as hepatocellular carcinoma, leiomyoma, sarcoma, sclerosed haemangioma and inflammatory pseudotumor may show identical features [3,4].

Therefore, the definitive diagnosis is based on typical histopathological and immunohistochemical features [5–7]. Herein we report two cases of SFT of the liver and discuss the relative diagnostic and therapeutic implications.

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http://dx.doi.org/10.1016/j.ijsu.2014.10.037
1743-9191/

2. Case report

2.1. Case 1

A 65-year-old man was accidentally found to have a large mass in the liver on an ultrasound carried out for an urological work up. An abdominal Magnetic Resonance Imaging (MRI) showed a voluminous multilobulated mass in the left liver (Fig. 1). The mass appeared to have hypersignal on T2 weighted images and hyposignal on T1 weighted images. After gadolinium injection, multiple septations were seen enhancing in the portal and late phases. The mass had avascular portions representing necrotic tissues. The rest of the liver parenchyma was homogeneous. The tumor compressed the left portal branch and was in contact with the inferior vena cava. Biopsy of the mass showed tumoral cells demonstrating spindle and fibroblast-like cells within a collagenous stroma. Immunohistochemistry disclosed CD34 and vimentin positivity supporting the diagnosis of SFT. Left hepatectomy was performed with total hepatic vascular exclusion. The middle hepatic vein was feed from the tumor and was preserved. Grossly, the tumor was a large encapsulated mass with well limited margins weighting 4.8 kgs. Histology revealed a fasciculated mass with absence of necrotic or hemorrhagic degeneration. On immunohistochemistry, the tumor cells were strongly positive for vimentin, CD34, CD99, Bcl-2 and they were negative for Epithelial Membrane Antigen (EMA), actin muscle, CD117, S-100, IgG4 and Dog-1. 2% of the nuclei were positive for Ki-67. The postoperative course was complicated by a pneumothorax that required percutaneous drainage and the patient was discharged home on postoperative day 22. At 12 months of follow up, the patient was well and he had no signs of recurrence.

2.2. Case 2

An 87-year-old woman with a past medical history of stage IV cardiac failure and bearer of a pacemaker was referred to our hepatobiliary unit because a liver mass was discovered on Computed Tomography (CT) scan which was performed for a slight elevation in liver function tests. Alpha fetoprotein was normal. CT scan showed a voluminous, heterogeneous, hypodense mass occupying almost the whole of the right liver, measuring 14.6 × 12.3 × 17 cm, compressing the inferior vena cava. After contrast injection, the mass showed clear contrast uptake with a pseudocapsule, and pseudonecrotic zones.

There was no invasion of the portal vessels (Fig. 2). A thoracic, cerebral and bone CT scan did not show any distant metastases. A biopsy showed fusiforms cells without atypia, morphologic and immunohistochemical features in favor of a hepatic solitary fibrous tumor. On immunohistochemistry, the tumor cells were strongly positive for CD34, they were negative for PS-100, actin muscle, CD117 and desmin. Ki-67 showed rare positive nuclei. The diagnosis of SFT was made but no further intervention was planned as the surgical risk was too high. At 10 months of follow up, the patient was still alive.

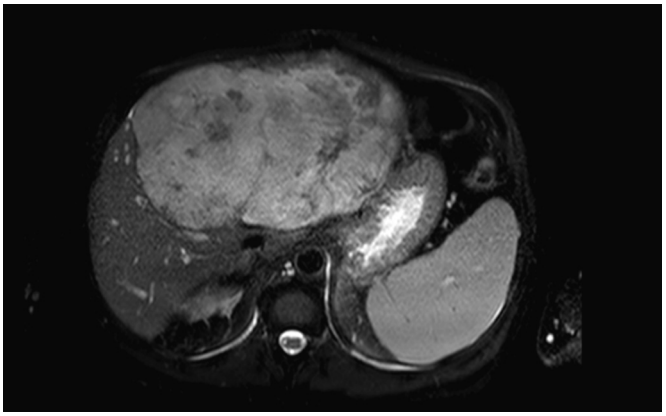

Fig. 1. MRI showing a large mass in the left liver.

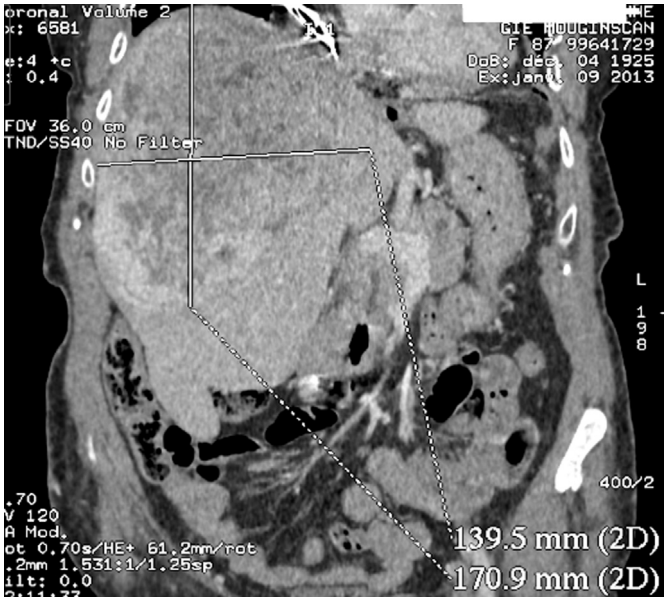

Fig. 2. CT Scan showing a large heterogeneous mass in the right lobe of the liver.

3. Discussion

We were able to find 59 cases of SFT that have been reported in the medical literature (Table 1). On reviewing all the reported literature, there does not seem to be a preference for these tumors to be present on one side of the liver over the other, with 29 cases in the right liver, 20 cases in the left liver and 10 cases were unknown. Female predominance exists with 70% women and 30% men. The differential diagnosis is very wide, and includes benign and malignant neoplasm [1–3]. As seen in our 2 cases, these tumors show heterogeneous enhancement on imaging, probably because of differential enhancement of the admixed cellular and collagenous components. However, imaging does not differentiate between benign and malignant tumors, as imaging features appear to overlap [3,4]. Symptoms usually appear late, and are mainly due to compression of nearby structures. Some patients presented with hypoglycemia, which can be explained by a Non-islet cell tumor hypoglycemia (NICTH), a syndrome related to paraneoplastic production of insulin-like growth factor (IGF). Some authors have associated the hypoglycemia to a poor prognostic sign. However, from the 5 patients who presented with symptoms of hypoglycemia, only one patient had a malignant tumor. So there does not seem to be enough criteria to correlate between hypoglycemia and malignancy [5,8,9]. A definitive diagnosis of SFT of the liver must therefore be made on the basis of histopathological and immunohistochemical studies with a high cellular proliferation of spindle cells arranged in a storiform pattern and with the immunohistochemical staining profile of CD34 (+), vimentin (+), Bcl-2 (+) and SM-actin (−). These characteristics differentiate SFTs from other liver tumors, such as primary hepatocellular carcinomas (CD34-negative), leiomyomas (smooth-muscle actin-positive and CD34-

Table 1
Cases with SFT found in the literature.

	Author	Year	Age/Gender	Malignancy	Hypoglycemia	Treatment	Liver	IH	F/up
1	Edmondson	1958	16/F	N	N	Excision	R	NA	24M
2	Edmondson	1958	NA	N	N	Excision	R	NA	NA
3	Nevius	1959	56/M	N	N	Radiation	R	NA	2D
4	Ishak	1959	56/M	N	N	Excision	L	NA	NA
5	Ishak	1976	62/F	N	N	Excision	L	NA	IOD
6	Kim	1983	27/F	N	N	Excision	L	NA	6M
7	Kottke-Marchant	1989	84/F	N	N	Excision	L	NA	29M
8	Kasano	1991	39/F	N	N	Excision	L	NA	53M
9	Barnoud	1996	50/M	N	N	Excision	R	CD34+	38M
10	Levine	1997	57/M	N	N	Excision	L	CD34+	NA
11	Guglielmi	1998	61/F	N	Y	Excision	R	CD34+	72M
12	Moran	1998	62/F	N	N	Excision	NA	CD34+	NA
13	Moran	1998	34/F	N	N	No Excision	NA	NA	AF
14	Moran	1998	57/F	N	N	Excision	NA	CD34+	NA
15	Moran	1998	32/M	N	N	Excision	NA	CD34+	NA
16	Moran	1998	68/F	N	N	Excision	NA	CD34+	2D
17	Moran	1998	83/F	N	N	Excision	R	CD34+	6D
18	Moran	1998	72/F	N	N	Excision	L	CD34+	12M
19	Moran	1998	62/F	N	N	Excision	L	CD34+	NA
20	Moran	1998	50/F	N	N	Excision	L	CD34+	NA
21	Fuksbrumer	2000	40/F	N	N	Excision	R	CD34+	NA
22	Fuksbrumer	2000	71/F	N	N	Excision	R	CD34+	NA
23	Fuksbrumer	2000	80/M	N	N	No Excision	R	CD34+	NA
24	Yilmaz	2000	25/F	Y + Metastasis	N	Excision	R	Vimentin+	6M
25	Lin	2001	75/M	N	N	Excision	R	CD34+	11M
26	Saint Marc	2002	69/F	N	N	Excision	R	CD34+	15M
27	Neeff	2004	63/F	N	N	Excision	L	CD34+	11M
28	Chithriki	2004	75/F	N	Y	Excision	R	CD34+	6M
29	Venarecci	2005	65/F	N	N	Excision	R	CD34+	30M
30	Ji	2006	46/F	N	N	Excision	R	CD34+	NA
31	Lehmann	2006	63/F	N	N	Excision	R	CD34+	96M
32	Nath	2006	65/F	N	N	Excision	R	CD34+	10M
33	Terkivatan	2006	74/M	N	N	Excision	L	CD34+	12M
34	Weitz	2007	NA	N	N	Excision	NA	NA	NA
35	Weitz	2007	NA	N	N	No Excision	NA	NA	NA
36	Weitz	2007	NA	N	N	No Excision	NA	NA	NA
37	Obuz	2007	52/M	N	N	Excision	L	CD34+	22M
38	Perini	2007	40/F	N	N	Excision	L	CD34+	49M
39	Chan	2007	70/M	Y + Metastasis	Y	Excision	R	CD34+	12M
40	Fama	2007	68/M	N	Y	Excision	R	CD34+	25M
41	Kwasky	2007	46/F	N	N	Excision	R	CD34+	NA
42	Kandpal	2007	45/F	N	N	No Excision	R	CD34+	NA
43	Korkolis	2008	82/F	N	N	Excision	L	CD34+	21M
44	Chen	2008	71/M	N	N	Excision	R	CD34+	9M
45	Novais	2008	34/F	N	N	Excision	R	CD34+	4Y
46	Taboada	2010	NA	NA	NA	NA	NA	NA	NA
47	Brochard	2010	54/M	Y + Metastasis	N	Excision	R	CD34+	6Y
48	Peng	2011	24/F	Y + Metastasis	N	Excision	R	CD34+	16M
49	Kesun	2011	59/M	N	N	Excision	L	CD34+	24M
50	Huanca	2011	52/NA	N	N	Excision	R	CD34+	NA
51	Arredondo	2011	NA	NA	NA	NA	NA	NA	NA
52	Patra	2012	34/F	N	N	Excision	L	CD34+	4Y
53	Radunz	2012	85/F	N	Y	Excision	L	CD34+	NA
54	Belga	2012	66/F	N	N	Excision	R	CD34+	2Y
55	Morris	2012	NA/F	N	N	Excision	R	CD34+	8M
56	Soussan	2013	64/M	N	N	Excision	L	CD34+	NA
57	Liu	2013	42/M	N	N	Excision	L	CD34+	NA
58	Jacob	2013	62/F	Y	N	Excision	L	CD34+	NA
59	**Present Case 1**	2013	65/M	N	N	Excision	L	CD34+	12M
60	**Present Case 2**	2013	87/F	N	N	No Excision	R	CD34+	10M
61	Song	2014	49/M	Y	N	Excision	R	CD34+	NA

F female; M male; Y yes; N no; Left; R right; M months; A years; NA not available; IH immunohistochemistry.

negative) and mesotheliomas (vimentin-positive, CD34-positive, cytokeratin-positive). However, the CD34 antibody is not specific for SFT diagnosis and can be positive in angiosarcomas and gastrointestinal stromal tumors, but it remains highly characteristic of SFT [8–11]. Biopsies might not be accurate to confirm a diagnosis because it is possible that benign and malignant tissues are present in the same lesion [11]. Fuksbrumer et al. [3] showed that malignant foci discovered on the resected specimen were not discovered on the initial needle core biopsy.

Therefore, a patient who only receive a needle core biopsy may have a tumor that contains undiagnosed malignant components. However biopsy might be helpful in cases where complete surgical resection is not possible or when a preoperative diagnosis is mandatory before surgery. Immunohistochemical study of the tissues taken from the biopsy can direct toward a diagnosis of SFT. These tumors might reach a very large size, and so at presentation, secondary vascular or bile duct compression may occur, as seen in some of the cases. The size and local invasion of these tumors to the

nearby vascular structures might lead to major hepatectomies, even with extensive resections with high morbidities and high mortalities [1,5,7,10]. The main concern in such cases is that SFT can develop malignant transformation [11]. It is possible that malignant and benign areas are present in the same lesion. The hypercellularity, nuclear atypia, necrosis and high mitotic activity (greater than 4 mitoses-ten high-power fields) are criteria used to diagnose malignancy. Larger tumors (>10 cm) are correlated with lower disease-free metastasis survival. Necrotic areas should alert the physician to the possibility of atypical features or malignant transformation, and biopsy may not identify a malignant component in the sample taken [8–11].

Six cases of malignant liver SFT, with 4 of them giving rise to metastases have been reported (Table 2). Brochard et al. [8] reported a 54-year-man with a right hepatic tumor that was surgically resected. Local recurrence occurred 6 years later, and it was resected. 2 years later, the patient developed metastasis to the cranial base, and he died a few months after [8]. Chan et al. [9] reported a 70-year-old man who presented with an SFT. A core needle biopsy was in favor of SFT. Resection was performed. The gross examination of the tumor revealed an SFT surrounded by a high grade fibrosarcoma. One year later, disseminated metastasis developed [9]. Peng et al. [12] presented a 24-year-old woman with a 30 cm mass in the right liver with skull base parietal metastasis. Extended right hepatectomy was performed and craniotomy was performed 13 days later. Pathological examination showed skull base metastases from a malignant solitary fibrous tumor of the liver. The tumor recurred in the residual liver 2 months after the operation, and lumbar vertebra metastasis 7 months after the operation paralyzed the patient. The patient died of uncontrolled tumor 16 months after the initial operation [12]. Yilmaz et al. [13] reported a 25-year-old woman who underwent right trisectionectomy for a liver mass. At follow-up one month later, the patient had severe back and waist pain. Metastatic osteoblastic lesions of the thoracic and lumbar vertebrae were identified on x-ray and CT scan. Biopsies of the left humerus and sternum revealed tumor destruction of bone by cells similar to that of the liver tumor. No follow up of the patient is available [13]. Jacob et al. [14] and Song et al. [15] reported 2 cases of malignant SFT of the liver without any distant metastasis. Surgical resection was performed; however no information regarding the follow up is available.

These 6 cases represent around 10% of SFT cases. The risk of malignancy is not extremely low, so it should always be kept in mind that SFT carries a real risk of malignant transformation [11–16].

In conclusion, it is difficult to differentiate benign from malignant tumors based only on imaging studies and biopsies. The natural history of the disease is unknown. These tumors can grow to large sizes and remain benign in nature. As these tumors can develop malignant transformation and they have the ability to spread and metastasize, surgical resection remains the optimal treatment, despite extensive surgery is needed to resect these tumors with clear margins. Complete surgical resection with clear margins remains the preferred treatment of choice. A correct interpretation should be done on the pathological and immunohistochemical markers for an appropriate diagnosis. However, it is difficult to determine the long term prognosis of this entity because of the small number of patients and the absence of long term follow up, which does not allow us to draw any meaningful conclusions regarding the clinical course of these tumors, particularly in those cases in which we observed atypical features.

Table 2
Summary of the 6 cases of malignant liver SFT.

Sex	3 Male; 3 Female
Age (years)	Mean 47; Range 24–70
Liver	5 Right; 1 Left
Metastasis	4 out of 6 patients (66%)
Hypoglycemia	1/6 (17%)
Treatment	6/6 Excision

Ethical approval

None required.

Funding

None.

Author contribution

Tarek Debs Radwan Kassir: writing.
Imed Ben Amor Francesco Martini: reviewed the pape.
Antonio Iannelli, Jean Gugenheim: conceptualized and designed the paper.

Conflict of interest

None.

References

[1] G. Vennarecci, G.M. Ettorre, L. Giovannelli, F. Del Nonno, L. Perracchio, P. Visca, V. Corazza, A. Vidiri, G. Visco, E. Santoro, Solitary fibrous tumor of the liver, J. Hepatobiliary Pancreat. Surg. 12 (4) (2005) 341–344.
[2] K. Sun, J.J. Lu, X.D. Teng, L.X. Ying, J.F. Wei, Solitary fibrous tumor of the liver: a case report, World J. Surg. Oncol. 9 (2011 Mar 29) 37.
[3] M.S. Fuksbrumer, D. Klimstra, D.M. Panicek, Solitary fibrous tumor of the liver: imaging findings, AJR Am. J. Roentgenol. 175 (6) (2000 Dec) 1683–1687.
[4] H. Kandpal, R. Sharma, S.D. Gupta, A. Kumar, Solitary fibrous tumour of the liver: a rare imaging diagnosis using MRI and diffusion-weighted imaging, Br. J. Radiol. 81 (972) (2008 Dec) e282–e286.
[5] F. Fama, Y. Le Bouc, G. Barrande, A. Villeneuve, M.G. Berry, R.R. Pidoto, O. Saint Marc, Solitary fibrous tumor of the liver with IGF-II-related hypoglycaemia. A case report, Langenbecks Arch. Surg. 393 (4) (2008 Jul) 611–616.
[6] S. Patra, M. Vij, K. Venugopal, M. Rela, Hepatic solitary fibrous tumor: report of a rare case, Indian J. Pathol. Microbiol. 55 (2) (2012 Apr-Jun) 236–238.
[7] P. Novais, C. Robles-Medranda, V.L. Pannain, D. Barbosa, B. Biccas, H. Fogaça, Solitary fibrous liver tumor: is surgical approach the best option? J. Gastrointestin. Liver Dis. 19 (1) (2010 Mar) 81–84.
[8] C. Brochard, S. Michalak, C. Aubé, C. Singeorzan, H.D. Fournier, L. Laccourreye, P. Calès, J. Boursier, A not so solitary fibrous tumor of the liver, Gastroenterol. Clin. Biol. 34 (12) (2010 Dec) 716–720.
[9] G. Chan, P.J. Horton, S. Thyssen, M. Lamarche, A. Nahal, D.J. Hill, E.B. Marliss, P. Metrakos, Malignant transformation of a solitary fibrous tumor of the liver and intractable hypoglycemia, J. Hepatobiliary Pancreat. Surg. 14 (6) (2007) 595–599.
[10] D.P. Korkolis, K. Apostolaki, C. Aggeli, G. Plataniotis, E. Gontikakis, D. Volanaki, M. Sebastiadou, D. Dimitroulopoulos, D. Xinopoulos, G.N. Zografos, P.P. Vassilopoulos, Solitary fibrous tumor of the liver expressing CD34 and vimentin: a case report, World J. Gastroenterol. 14 (40) (2008 Oct 28) 6261–6264.
[11] J.J. Chen, S.L. Ong, C. Richards, G. Garcea, C. Pollard, D. Berry, A. Dennison, Inaccuracy of fine-needle biopsy in the diagnosis of solitary fibrous tumor of the liver, Asian J. Surg. 31 (4) (2008 Oct) 195–198.
[12] L. Peng, Y. Liu, Y. Ai, Z. Liu, Y. He, Q. Liu, Skull base metastases from a malignant solitary fibrous tumor of the liver. A case report and literature review, Diagn. Pathol. 6 (2011 Dec 22) 127.
[13] S. Yilmaz, V. Kirimlioglu, E. Ertas, F. Hilmioglu, B. Yildirim, D. Katz, B. Mizrak, Giant solitary fibrous tumor of the liver with metastasis to the skeletal system successfully treated with trisegmentectomy, Dig. Dis. Sci. 45 (1) (2000 Jan) 168–174.
[14] M. Jakob, M. Schneider, I. Hoeller, U. Laffer, R. Kaderli, Malignant solitary fibrous tumor involving the liver, World J. Gastroenterol. 19 (21) (2013 Jun 7) 3354–3357.
[15] L. Song, W. Zhang, Y. Zhang, (18)F-FDG PET/CT imaging of malignant hepatic solitary fibrous tumor, Clin. Nucl. Med. 39 (7) (2014 Jul) 662–664.
[16] D.S. Nath, A.D. Rutzick, T.D. Sielaff, Solitary fibrous tumor of the liver, AJR Am. J. Roentgenol. 187 (2) (2006 Aug) W187–W190.